CASE REPORT

Hypoplastic Left Heart Syndrome

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ABSTRACT

Hypoplastic left heart syndrome (HLHS) may result from errors in the early stages of cardiac development. HLHS represents a variety of cardiac malformations that include hypoplasia of the left ventricle, hypoplasia of ascending aorta and hypoplasia or atresia of the aortic and mitral valves. By the help of improved resolution of advanced sonography equipments a radiologist can diagnose the spectrum of HLHS. HLHS causes increase in neonatal morbidity and mortality. Hence there is a need for prenatal diagnosis of HLHS with ultrasound to detect cardiac anomalies. Prenatal diagnosis of this congenital cardiac anomaly also allows families to prepare for a child with a life-threatening defect by consultation with the multidisciplinary team for short- and long-term prognosis.

INTRODUCTION

Hypoplastic left heart syndrome (HLHS) is mostly a lethal condition. Spectrum of HLHS includes hypoplasia of the left ventricle and ascending aorta and hypoplasia or atresia of the aortic and mitral valves. The great vessels are normally positioned in HLHS. HLHS is the fourth most common cardiac malformation to manifest in the 1st year of life behind ventricular septal defect, d-transposition of the great arteries, and tetralogy of Fallot[1]. HLHS has a reported prevalence of 0.2 per 1,000 live births and occurs twice as often in boys as in girls[2,3]. Left untreated, HLHS is invariably lethal and is responsible for 25% of early cardiac deaths in neonates[4,5]. However, the recent evolution of palliative surgical procedures has increased the survival rate in children with these malformations.

CASE PRESENTATION

A 29 year-old primigravida of Indian origin, 34 weeks of amenorrhea presented to obstetrical outpatient department for routine antenatal check-up. The patient's obstetrical history was normal with no significant past medical or family history. There was no significant history of drug intake. Her menstrual cycles were regular. Her sonography findings were small thick wall left ventricle with small ascending aorta and normal pulmonary artery with ratio of aorta to pulmonary artery of 0.6, enlarged right heart chambers with reduced flow across mitral valve with normal left atrium and heart rate was 222 beats/min with regular rhythm.

DISCUSSION

HLHS is a complex combination of cardiac malformations that probably results from multiple developmental errors in the early stages of cardiogenesis. As fetal circulation allows normal growth and development of the fetus, the ductus arteriosus and elevation of pulmonary arterial pressures in the early postnatal period leads to poor perfusion of the coronary, cerebral and systemic circulations leading to severe congestive heart failure. If left untreated it may lead to premature death. A variety of chest radiographic findings are seen in patients with HLHS and multiple innovative surgical procedures are now available to prolong the lives of these patients. An awareness and understanding of the spectrum of radiographic findings in HLHS will help radiologist to better assist their colleagues in cardiology and cardiovascular surgery in caring for patients with this pathologic condition.

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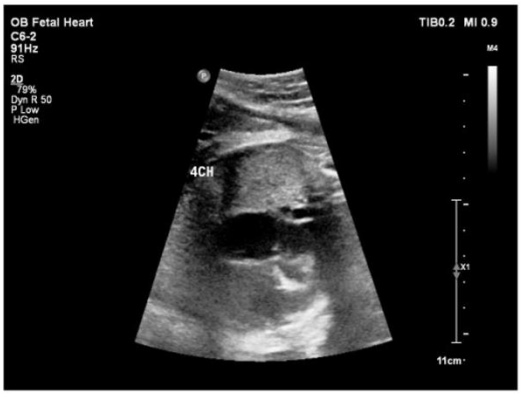
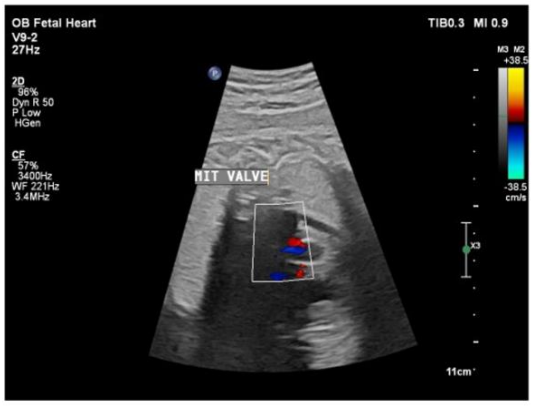


Figure 1: A- 4 chamber heart view shows small left ventricle with enlarged right chambers.



B- Decreased flow across mitral valve.

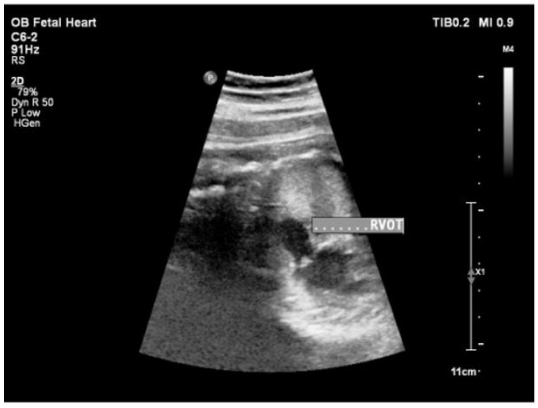
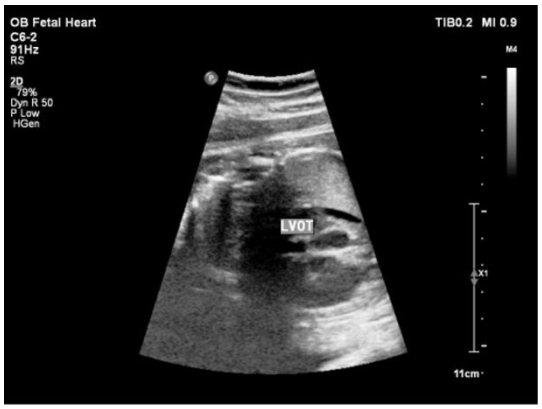


Figure 2 : A- Normal RVOT



B- Narrow LVOT

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